Case Report

Abdominal mass mimicking a leiomyoma: Malignant uterine solitary fibrous tumor

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Introduction

Uterine sarcomas encompass a broad spectrum of neoplasms that range from pure mesenchymal tumors and endometrial stromal tumors to mixed epithelial/stromal tumors. Although several classification systems are available, they are all based upon the World Health Organization histologic classification system of uterine mesenchymal tumors. Formerly believed to be restricted to the pleura, solitary fibrous tumors (SFTs) have been increasingly recognized in extrapulmonary sites (Morimitsu et al., 2000; Vallat-Decouvelare et al., 1998). In the female genital tract they are extremely rare and there are only scarce reports in the literature (Berzal-Cantalejo et al., 2005). Typical histologic features of SFTs include haphazardly arranged spindle cells, alternating hypocellular and hypercellular areas, collagenous stroma, and thin-walled branching blood vessels (Morimitsu et al., 2000; Vallat-Decouvelare et al., 1998; Berzal-Cantalejo et al., 2005). Although myxoid change is not an uncommon finding in SFTs, those with a prominent myxoid stroma are quite rare (De Saint Aubain Somerhausen et al., 1999; Lau et al., 2009).

Case report

A 30-year-old woman presented with diffuse abdominal pain and menorrhagia for the last four months. Physical examination revealed

an enlarged uterus (17 weeks size). The transvaginal ultrasound demonstrated a uterine intramural/subserosal nodule compatible with leiomyoma. Laboratory data was unremarkable. The patient underwent transabdominal myomectomy and the excision of the tumor (with a jelly-like gross appearance) was fractionated and incomplete. Macroscopic examination showed a myxoid fragmented mass. Microscopically, the neoplasm was characterized as a mesenchymal tumor, with spindle cells on a myxoid stroma with alternating hyper- and hypocellular areas (Fig. 1A). Hemangiopericytoma-like vessels were also described (Fig. 1D). Mitotic rate was 4 per 10 high power fields (Fig. 1C). There were no necrotic areas. Immunohistochemical analysis showed that the neoplastic cells were diffusely immunoreactive for CD 34, CD 99, and bcl2 (Fig. 2); they were negative for CAM 5.2, HHF-35, smooth muscle actin, CD 10, S-100 and desmin. Histological and immunohistochemical features suggested a myxoid malignant SFT of the uterus. Brain magnetic resonance imaging (MRI), thoracic, abdominal and pelvic computed tomography (CT) scans excluded the presence of extra-pelvic neoplastic sites and metastasis. Hysterectomy with bilateral adnexal conservation was performed afterwards. Histological examination showed persistence of the myxoid SFT previously described, with infiltrative growth towards the myometrium. Currently the patient is being followed (50 months after the surgery) and all radiographic data shows no evidence of disease.

Discussion

SFTs are uncommon neoplasms. Initially identified as a distinct pleural lesion, SFTs are tumors of ubiquitous location and they were described at various extrapleural sites. In the female tract, SFT is extremely uncommon (Berzal-Cantalejo et al., 2005). Usually, patients range from 20 to 70 years. Symptoms, if present, include painless swelling, menorrhagia, flank pain or pelvic fullness. One uterine SFT was associated with severe hypoglycemia due to insulin-like growth factor II production (Vallat-Decouvelare et al., 1998). Myxoid change is a common, though generally focal finding in SFTs. In contrast, SFTs exhibiting a predominantly myxoid stroma are rarely encountered. The histologic features of myxoid solitary fibrous tumor are relatively nonspecific and include the presence of abundant, pale myxoid matrix, prominent, branching, staghorn-like (hemangiopericytoma-like) blood vessels, and a “patternless pattern” growth of spindled cells, with alternating hyper- and hypocellular areas (De Saint Aubain Somerhausen et al., 1999; Lau et al., 2009). Similar to conventional SFT, by immunohistochemistry, the myxoid variant commonly expresses CD34, CD99, and bcl-2, and is typically negative for keratin, epithelial membrane antigen (EMA), S-100 protein, and muscle-associated antigens (Lau et al., 2009).

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Fig. 1. A — Hypercellular areas alternating with myxoid dominant areas (H&E stain, 40×). B — Perivascular hyalinization (H&E stain, 200×). C — Mitotic activity (black arrows, H&E stain, 400×). D — Collagenous bundles, rosettes and hemangiopericytoma-like vascular pattern (H&E stain, 200×).

The differential diagnosis of myxoid solitary fibrous tumor is rather broad, and includes the myxoid variant of other spindle cell neoplasms such as low grade fibrosarcoma, fibrosarcomas, synovial sarcomas, leiomyomas, leiomyosarcomas, liposarcomas, and nerve sheath tumors; all of these neoplasms are generally CD34 negative. Low-grade fibrosarcoma contains myxoid areas and cytologically, spindle cells of this tumor adopt a characteristic whorled or swirling growth pattern not seen in myxoid SFT (Lau et al., 2009). The presence of multinodular growth with fibrous septa, cells with atypical hyperchromatic nuclei and

mucin containing pseudolipoblasts are diagnostic hallmarks of myxofibrosarcomas (Lau et al., 2009). Myxoid synovial sarcoma can be distinguished from myxoid SFT because it is usually immunoreactive for keratin and epithelial membrane antigen (EMA) (Lau et al., 2009). Myxoid leiomyoma and leiomyosarcomas typically show positivity for desmin, actin and smooth muscle actin, which are generally not detected in myxoid SFT (Table 1). Myxoid liposarcoma is separable from myxoid SFT by the presence of lipoblasts, an arborizing capillary vasculature and pools of stromal mucine. Nerve sheath tumors and

Fig. 2. Immunohistochemistry of the spindle cells revealed the expression of CD 34 and BCL2.

Table 1

Histology and staining patterns of leiomyomas, leiomyosarcomas and SFTs.

	Histology	Immunohistochemistry
Leiomyoma	Fascicles of elongated spindle cells with ovoid, “cigar-shaped” nuclei and eosinophilic cytoplasm	Desmin (+) h-Caldesmon (+) HDAC8 (+) Smooth muscle actin (+) CD10 (+/–)
Leiomyosarcoma	Hypercellular, fascicular growth; areas of hypocellularity may be present Elongated spindle cells Tumor cell necrosis Mitotic index $\geq 10/10$ HPF Diffuse moderate to severe cytologic atypia	Desmin (+) h-Caldesmon (+) Smooth muscle actin (+)
SFT	Patternless growth with alternating hyper- and hypocellular areas Abundant pale myxoid matrix Prominent, branching, staghorn-like blood vessels (hemangiopericytoma-like)	Desmin (–) Smooth muscle actin (–) h-Caldesmon (–) CD 10 (–) CD 34 (+) bcl2 (+) Vimentin (+) CD 99 (+/–)

especially malignant peripheral nerve sheath tumor are S-100 protein positive in one half of the cases (myxoid SFTs are generally negative for this marker).

The biological behavior of extrapleural SFT has been regarded as unpredictable (Morimitsu et al., 2000; Vallat-Decouvelare et al., 1998; Berzal-Cantalejo et al., 2005; De Saint Aubain Somerhausen et

al., 1999; Lau et al., 2009; Hasegawa et al., 1999). Some tumors with atypical histological features (such as, nuclear atypia, infiltrative margins, high cellularity, increased mytotic rate and foci of hemorrhage and necrosis) did not behave aggressively and other tumors without any atypical histologic features developed local recurrence or distant metastasis (Morimitsu et al., 2000; De Saint Aubain Somerhausen et al., 1999; Hasegawa et al., 1999). Due to the lack of clinical information concerning the behavior of myxoid SFT and the histologic findings in this particular case, extended follow-up is required. After our research (English literature), we found no other case of a malignant myxoid SFT of the uterus with a mainly intramural location.

Conflict of interest statement

The authors declare that there is no conflict of interest.

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